Heart Disease in Children*

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Fifteen years ago, a diagnosis of "congenital heart disease" might have been considered adequate. Today, all pediatricians, radiologists, surgeons, and other physicians would insist on more precise anatomic and physiologic information.

The reason for this striking change is shown in table 1. All the lesions listed can be corrected by surgery. They are, then, potentially curable diseases. Almost all of the common congenital heart diseases are in this group. In addition, there are many active programs in clinical research centers searching for satisfactory correction of still other lesions, such as complete transposition of the great vessels (the second most common cyanotic heart disease), Ebstein's anomaly, and single ventricle. Even among those conditions that cannot be cured or corrected, ameliorative procedures are now available. The Blalock operation for tetralogy of Fallot is such a procedure. In this operation, the subclavian artery is used as a shunt vessel to carry blood from the systemic circuit into the pulmonary arterial tree. Another is the Glenn procedure, a superior vena cava-pulmonary artery anastomosis.

Anomalies of the cardiovascular system are a common cause of disease in children. Approximately six of every one thousand live births have some form of congenital heart disease. The prognosis of many of these conditions is very poor. About 90% of infants born with complete transposition of the great vessels succumb within the first year of life. Consequently, early and accurate diagnosis is essential.

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Classification of Congenital Heart Disease

Classifications based on either anatomic or physiologic pathology are helpful in this regard, and many have been proposed. Because most of the observable clinical abnormalities are reflections of pathologic physiology, the latter approach is more useful.

The simplest classification of this sort is based on the presence or absence of visible cyanosis (table 2A). Patients with desaturated hemoglobin in the peripheral arterial circulation (that is, with right-to-left shunt of blood), as in tetralogy of Fallot or complete transposition of the great vessels, are separated from those with

TABLE 1

Some Cardiac Lesions that can be Corrected

Ventricular septal defect	Subvalvular aortic stenosis
Atrial septal defect	Tetralogy of Fallot
Atrioventricularis communis	The grand trilogy
Patent ductus arteriosus	Total anomalous pulmonary venous connection
Aortico-pulmonary window	Partial anomalous pulmonary venous connection
Coronary artery fistula	Pulmonary valvular stenosis
Ruptured sinus of Valsalva	Aortic ring
Coarctation of the aorta	Cardiac tumors
Aortic stenosis	Congenital ventricular aneurysm
Supravalvular aortic stenosis	Congenital mitral stenosis

TABLE 2

Classifications of Congenital Cardiac Lesions

A Clinical Classification	B Roentgenologic Classification
Acyanotic	*Increased pulmonary arterial vasculature*
Left to right shunts	Left to right shunts
Stenotic lesions: right sided	Admixture lesions
left sided	*Normal pulmonary arterial vasculature*
Myocardial lesions	Stenotic lesions: left sided
Some pulmonary venous obstructive lesions	right sided
Cyanotic	Myocardial lesions
Pulmonary obstruction with right to left shunt	*Decreased pulmonary arterial vasculature*
Admixture lesions	Pulmonary obstruction with right to left shunt
Some pulmonary venous obstructive lesions	*Increased pulmonary venous vasculature*
	Pulmonary venous obstructive lesions

a left-to-right shunt (such as ventricular septal defect) or those with cardiac conditions without shunt (e.g., coarctation of the aorta).

This classification has inherent disadvantages, however. Many infants and children with conditions characterized by right-to-left shunts have an insufficient quantity of desaturated hemoglobin in the peripheral circulation to be observably blue. About 5 g% of desaturated hemoglobin is required for visible cyanosis, and this is dependent upon a number of factors, such as skin pigmentation. In addition, cyanosis may result from extensive pneumonia, and other pulmonary conditions. For these reasons, a classification (table 2B), based on radiologic observation of the pulmonary blood vessels, was proposed a number of years ago (Lester, Gedgaudas, and Rigler, 1958). This method, too, is based on physiologic considerations. Congenital cardiac conditions are separated into groups dependent upon the pulmonary vascular blood flow.

Decreased Pulmonary Arterial Vasculature

Patients with a decrease in pulmonary arterial vasculature must have both an obstruction of pulmonary flow (such as pulmonary stenosis or right ventricular infundibular obstruction) and a right-to-left shunt of blood (e.g., through a ventricular septal defect). Tetralogy of Fallot is an example of this, and is defined as infundibular stenosis with ventricular septal defect and right-to-left shunt. Other lesions falling into this group are shown in table 3. Patients with these lesions show arterial O_2 desaturation and may be visibly cyanotic. Differential diagnoses within this or other groups may be accomplished by further roentgen observations.

Increased Pulmonary Arterial Vasculature

Patients showing an increase in pulmonary arterial vasculature fall into one of two sub-groups. By far, the more common is the left-to-right shunt series. These include intracardiac left-to-right shunts, such as ventricular septal defect, and atrial septal defect.

TABLE 3

Decreased Pulmonary Arterial Vasculature (Cyanotic)

- Tetralogy of Fallot
- The grand trilogy
- Pulmonary atresia
- "Pseudotruncus arteriosus"
- Tricuspid atresia
- Ebstein's anomaly (with right to left atrial shunt)
- Primary pulmonary hypertension with patent foramen ovale
- Pulmonary arterial hypoplasia or coarctation (may be unilateral)
- Origin of both great vessels from right ventricle with pulmonary stenosis
- Complete transposition of the great vessels with pulmonary stenosis or atresia
- Truncus arteriosus, type IV

Such patients are, of course, fully saturated. In this series, increased pulmonary vasculature is associated with an enlarged, undivided pulmonary artery, and with an aorta that appears normal or somewhat small, and is relatively hypopulsatile. Extracardiac left-to-right shunts include patent ductus arteriosus and aortico-pulmonary window. In these patients, the aorta appears hyperpulsatile and may be enlarged. In addition, extracardiac to intracardiac left-to-right shunts may be seen, such as aneurysm of a sinus of Valsalva with fistula formation and coronary artery fistula (table 4).

In addition, patients with admixture lesions, where both a left-to-right shunt and a right-to-left shunt are essential for maintaining life, usually show increased pulmonary arterial vasculature. In truncus arteriosus, for example, the blood flow from both the left and right ventricles enters a common trunk, from which the flow into the systemic and the pulmonary circulations arises. In complete transposition of the great vessels, too, there is invariably shunting in both directions. These patients are, of course, desaturated, and may be visibly blue. In the admixture group there is frequently (although not always) some degree of malposition of the great vessels. As a result, although the pulmonary arterial vasculature is increased on the roent-

TABLE 4

Increased Pulmonary Arterial Vasculature (Acyanotic)

Left to Right Shunts

- Ventricular septal defect
- Atrial septal defect
- Atrioventricularis communis
- Left ventricular-right atrial communication
- Patent ductus arteriosus
- Aortico-pulmonary window
- Ruptured sinus of Valsalva
- Coronary artery fistula to right atrium, right ventricle or pulmonary artery
- Partial anomalous pulmonary venous connection
- Lutembacher's syndrome
- Corrected transposition with left to right shunt

TABLE 5

Increased Pulmonary Arterial Vasculature (Cyanotic)

Admixture Lesions

- Complete transposition of the great vessels
- Origin of both great vessels from right ventricle without pulmonary stenosis
- Other forms of partial transposition of the great vessels
- Truncus arteriosus, types I, II, III
- Single ventricle
- Cor biloculare
- Total anomalous pulmonary venous connection
- Tricuspid atresia with transposition of the great vessels

Shunt with High Pulmonary Resistance

- "Eisenmenger complex" and "Eisenmenger physiology"

genograms, the area of the undivided pulmonary artery may appear flat or concave. This is not because the pulmonary artery is small, but rather because it is malpositioned (table 5).

Normal Pulmonary Vasculature

When the pulmonary arterial vasculature is normal, the lesion is of a variety not associated with shunt mechanisms. Of course, a shunt defect not physiologically significant will also show no abnormality of pulmonary vasculature. These lesions too can be sub-grouped into left-sided ones (coarctation of the aorta), right-sided (isolated pulmonary valvular stenosis), and more generalized ones (Von Gierke's Disease) (table 6).

Pulmonary Venous Obstruction

In addition, there is a series of relatively rare but important diseases, characterized by pulmonary venous obstruction. These lesions can be recognized by a reticular pattern in the lung fields that represents, not engorged pulmonary arterial vessels, but engorged venous vessels and lymphatics. These patients also may show repeated bouts of pulmonary edema. Such conditions as cor triatriatum and total anomalous pulmonary venous connection to the portal venous system fall into this group (table 7). It should be noted that the more common varieties of total anomalous pulmonary venous connection are admixture lesions, not pulmonary venous obstructive lesions.

Choice of Radiologic Procedure

Although the status of the pulmonary arterial and venous vasculature is best defined in the frontal roentgenogram, other findings are of particular help in the differential diagnosis of conditions within the major groupings. These include the size and pulsations of the aorta, the pulsations of the pulmonary arterial vessels themselves, the size of the left atrium, and the size of the ventricles. For this reason, the frontal view chest roentgenogram is not a satisfactory examination of the heart. Rather, a series of films and a brief and appropriate fluoroscopic examination are required for full evaluation.

TABLE 6

Normal Pulmonary Arterial Vasculature (Acyanotic)

Coarctation of aorta	Von Gierke's disease
Aortic stenosis or insufficiency	Hurler's disease and other metabolic heart disease
Aortic ring	Sickle cell disease and other anemias
Endocardial fibroelastosis	Congenital ventricular aneurysm
Cor triatriatum	Anomalous origin of a coronary artery
Pulmonary valvular stenosis	Coronary artery fistula to left side of heart
Friedreich's ataxia heart disease	Complete heart block
Marfan's disease	Cardiac tumors
	Hypertensive heart disease

TABLE 7

Increased Pulmonary Venous Vasculature

- Stenosis or atresia of pulmonary veins
- Cor triatriatum
- Mitral atresia
- Mitral stenosis
- Anomalous pulmonary venous connection, through stenotic channels
- Anomalous pulmonary venous connection to the portal venous system
- Aortic atresia
- Severe aortic stenosis
- Severe coarctation of the aorta
- Complete interruption of the aortic arch

TABLE 8

Initial Cardiac Films

(All Films at 6 Foot Target-Film Distance—Upright)

- Postero-anterior*
- Lateral*
- Right anterior oblique, 45°*
- Left anterior oblique, 60°

* Thick barium in esophagus.

Additional Films

- Supine antero-posterior at 30 inch distance—for left atrium
- Roentgen kymograms—for valve calcification, pulsations
- Cinefluorographic strips—for pulsations, calcifications
- Planigrams—for valve calcification

The routine cardiac films most suitable for differential diagnosis are listed in table 8.

The status of fluoroscopic examination has been brought into question in recent years. This is a result of misunderstanding the use of the fluoroscope in these conditions. It is feckless to use the fluoroscopic examination for evaluating factors which can be determined better from the roentgenograms. On the other hand, dynamic characteristics, such as the pulsations of the undivided pulmonary artery and its major branches and the pulsations of the aorta, can be evaluated only by fluoroscopy. For this purpose, image intensification fluoroscopy, rather than a conventional fluoroscope, is essential.

Evaluation of Radiographic and Fluoroscopic Findings

An order of significance of radiographic and fluoroscopic findings can be derived. Some features, such as the status of the pulmonary arterial vasculature, are well evaluated radiographically and are consequently of great significance. Others, such as chamber enlargement with the exception of left atrial enlargement, are poorly evaluated radiographically and other methods are more satisfactory. An order of significance is indicated in table 9.

Utilizing simple roentgen methods (radiographic films and image intensification fluoroscopy) in association with the other simple diagnostic methods (physical examination, history, and

electrocardiography), a firm clinical diagnosis can be established in more than 85% of the patients. However, further information is usually needed. A decision to attempt surgical correction depends upon precise knowledge of the physiologic effect of the anatomic defect. For example, a patient with ventricular septal defect, having only a modest increase in pulmonary arterial pressure, represents an excellent surgical risk. Appropriate repair should result in complete correction of the condition. On the other hand, a patient having very high pulmonary resistance may be a poor risk, and surgery may be contra-indicated. This sort of data is best established by cardiac catheterization.

Selective Angiocardiography

Surgical correction also is dependent upon a knowledge of the precise anatomic pathology. In tetralogy of Fallot, for example, the degree of infundibular obstruction, the length of the hypertrophied crista supraventricularis, the size of the ventricular septal defect, the degree of pulmonary valvular obstruction, and the degree of overriding of the aorta are all of great importance. This sort of precise anatomic information requires selective angiocardiography.

Generally, venous angiocardiography (injection of a bolus of contrast material into a peripheral vein with filming of the heart) is unsatisfactory because of overlapping structures and a poor concentration of contrast material at the point of interest. Selective study, with a catheter placed in relation to the expected area of maximum importance, is much superior and should replace the cruder methods. In order to accomplish this, the conventional studies must be used in order to determine what information is to be obtained. Right-sided selective angiocardiography is most satisfactory in a large variety of conditions. These include tetralogy of Fallot and pulmonary stenosis (table 10). Injection into the left side of the heart is required in others (table 11). In extracardiac conditions involving the aorta, selective injection above the aortic valve, or other locations within the aorta, may be necessary (table 12).

TABLE 9

Radiographic and Fluoroscopic Findings in Order of Significance

- Pulmonary arterial vasculature
  - a. Normal, increased or decreased on radiographs
  - b. Degree and character of pulsations at fluoroscopy
- Pulmonary venous vasculature and pulmonary lymphatics
- Size and pulsations of undivided pulmonary artery
- Size and pulsations of aorta
- Heart size
- Chamber enlargement
  - a. Left atrium
  - b. Right ventricle
  - c. Left ventricle
  - d. Right atrium

TABLE 10

Some Indications for Right Sided Angiocardiography

- Congenital heart disease with decreased pulmonary arterial vasculature
- Congenital heart disease with increased pulmonary arterial vasculature
  - a. Admixture lesions
  - b. Bidirectional shunt
- Anatomic definition of pulmonary valvular and/or infundibular stenosis
- Anomalies of the pulmonary vascular tree
- Inability to enter the pulmonary artery at cardiac catheterization
- Congenital heart disease with increased pulmonary venous vasculature
- Differential diagnosis between cardiac dilatation and pericardial effusion
- Cardiac tumors
- Certain pericardiac tumors

TABLE 11

Some Indications for Left Sided Angiocardiography

- Evaluation of mitral competance in acquired heart disease
- Evaluation of mitral competance following surgery for mitral disease
- Subvalvular aortic stenosis
- Small intracardiac and extracardiac left to right shunts with questionable findings at right heart catheterization; anatomic definition of such lesions
- Balanced septal defects
- Patent ductus arteriosus associated with pulmonary insufficiency or with ventricular septal defect
- Certain admixture lesions
- Evaluation of left side of heart in complex congenital anomalies

TABLE 12

Some Indications for Thoracic Aortography

- Coarctation of the aorta
- Patent ductus arteriosus
- Aortico-pulmonary window
- Coronary artery fistula
- Anomalous coronary artery
- Ruptured sinus of Valsalva
- Aortic stenosis and/or insufficiency
- Supravalvular aortic stenosis
- Aortic ring
- Aneurysm of the aorta

Summary

1. The importance of precise anatomic and physiologic diagnosis in congenital heart disease has been emphasized in recent years, because of the increasing possibilities of surgical correction.

2. The x-ray examination is a powerful tool in the evaluation of patients with congenital heart disease. A method of evaluation is presented, using conventional roentgenographic and fluoroscopic examination along with other simple clinical methods. This approach is relatively simple, can be carried out as an office procedure, and yields a firm clinical diagnosis in the majority of cases.

3. Cardiac catheterization and selective angiocardiography may be necessary to give precise information on physiological abnormalities and anatomic defects. These techniques require expensive equipment and a team of physicians.

4. The challenge of accurate diagnosis in these common anomalies is great. The potential reward, in terms of restoring a normal life span, is also great. An aggressive approach to a correct diagnosis is urged.

Reference

LESTER, R. G., E. GEDGAUDAS, AND L. G. RIGLER. Method of radiologic diagnosis of congenital heart disease in children. *J. Am. Med. Assoc.* 166: 439–443, 1958.